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Fig. Ia Control radiograph.



Fig. Ib Full length intravenous urogram.

CASE PRESENTATION

A 56-year-old man was being investigated for microhaematuria. He was asymptomatic, and had a background history of hypertension and diabetes mellitus. His serum creatinine was normal but creatinine clearance was 24 ml/min. What does the intravenous urogram (IVU) (Figs. 1a-b) show? What other imaging modalities would be useful? Department of Urology Singapore General Hospital Outram Road Singapore 169608

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Fig. 2a Enhanced CT scan taken at the upper pole level shows the close proximity of the upper pole calyx (the preferred percutaneous puncture site) to the tip of the right lobe of liver (arrowheads). Both kidneys are hydronephrotic.



Fig. 2b Enhanced CT scan taken at hilar level shows multiple bilateral renal pelvic and calyceal stones in moderately hydronephrotic kidneys.



Fig. 2c Enhanced CT scan taken at the lower pole level shows the bridging renal tissue (isthmus) (arrow) joining the lower poles of both kidneys.



Fig. 3 Unenhanced CT scan taken with the patient in the prone position shows the needle in the upper pole calyx of the left kidney. The puncture site is between the 11th and 12th rib and the pleura is unlikely to be affected.

IMAGE INTERPRETATION

The control radiograph (Fig. 1a) shows multiple stones projected over the renal outlines. IVU (Fig. 1b) confirms that the stones are located within bilateral moderately-severe hydronephrotic kidneys. Both kidneys are fused at their lower poles, with medial rotation of their axes. Computed tomography (CT) of the kidneys (Figs. 2a-c) was performed to accurately delineate the calyceal systems in relation to the other organs, such as the colon, spleen and liver. The spleen was very closely related to the upper pole calyx (Fig. 2a), which is the preferred calyx for puncture in percutaneous nephrolithotripsy (PCNL).

DIAGNOSIS

Hydronephrotic horseshoe kidneys with multiple calculi.

CLINICAL COURSE

The patient underwent a CT-guided percutaneous puncture of the left upper pole in order to avoid injury to the spleen (Fig. 3). This was followed by an uneventful PCNL in the operating theatre. The post-operative radiograph showed complete stone clearance for the left-sided urinary system (Fig. 4a). The right collecting system was similarly treated two months later. Only the right renal pelvic stones were cleared during the first session of the PCNL (Fig. 4b). A secondary PCNL to achieve stone clearance status was planned. However, the patient was not agreeable and it was decided that extracorporeal shock wave lithotripsy (ESWL) would be employed for the remnant fragments.

DISCUSSION

Horseshoe kidney is the most common fusion anomaly, with an incidence of 0.25% in the general population⁽¹⁾. As a result of the fusion, there is failure of ascent and normal lateral rotation of the kidneys. The collecting system is displaced anteriorly, and the ureters arise high from the renal pelvis and descend anterior to the isthmus. There is impaired drainage of the collecting system, hence 17% - 21% of horseshoe kidney patients have stones⁽¹⁾. Infection, stone formation and subsequent renal impairment are therefore common sequelae in untreated horseshoe kidneys.

The initial work-up is geared towards identification of stone-forming bacteriae, such as *Proteus mirabilis*, and the assessment of renal function. The first clue to the diagnosis may be seen on the abdominal radiograph. Medial rotation of the both renal long axes would suggest the presence of horseshoe kidneys. An IVU or renal ultrasonography can be used to confirm the medial rotation of the renal axes, which results in fusion of the kidneys at their lower poles. The renal isthmus may be no more than a fibrous band or consist



Fig. 4a Radiograph taken post-left PNL shows complete left stone clearance.



Fig. 4b Radiograph taken post-first session right PCNL shows a nephrostomy tube in-situ. The access track is thus maintained for secondary PCNL aimed at stone clearance.

of "normal" renal parenchyma. In the latter instance, the isthmuses will opacify together with the rest of the kidney during the nephrographic phase of the IVU study. On ultrasonography, the relative hypoechogeneity of normal renal parenchyma will be seen to continue across the midline, anterior to the aorta, and merging with the opposite kidney.

CT scan can also elegantly demonstrate all the salient features of a horseshoe kidney, including the pre-aortic isthmus at L4/5 level and anterior displacement of the ureters which drape over the isthmus. Some authors have also reported the utility of the CT in detecting the interposition of colon, liver or spleen between the skin surface and the kidneys⁽²⁾. This imaging finding may alter the treatment plans. For example, as in this case, due to the proximity of the spleen to the upper renal pole, a CT-guided percutaneous puncture was chosen (Fig. 3) in preference to fluoroscopic-guided puncture in the operating theatre.

Treatment of stones in such renal anomalies is guided by the anatomy and stone burden. A patient with low stone burden, less than 2 cm in size, may be amenable to ESWL treatment. As the collecting system is displaced anteriorly, there is always a concern for the adequacy of drainage after lithotripsy. Hence, ESWL treatment in the prone position is preferred^(3,4). Stone-free rates after ESWL treatment of such stones are in the range of 50% - 78% ⁽⁵⁻⁸⁾. This is comparable to stone-free rates of 85% - 90% for renal stones less than 2 cm in normal renal system⁽⁹⁾. Several authors have also alluded to the high rate (21% - 50%)^(7,8,10) of "clinically-insignificant fragments" after ESWL. This may be due to poor localisation or poor drainage despite good fragmentation due to the anomalous renal anatomy.

Percutaneous nephrolithotripsy is a good alternative for treatment of stones larger than 2 cm in anomalous systems. CT-guided drainage is also indicated for patients with structural abnormalities, such as incisional hernia from previous open pyelolithotomy (Fig. 5). For simple malrotation, usual fluoroscopic-guided puncture is adequate if the CT shows that the adjacent structures are not at risk (Fig. 6a-b). For horseshoe kidneys, the upper pole calyx is the calyx of choice for puncture because it occupies the most posterior position within the kidney. The risk of bleeding is similar to that of kidneys with normal anatomy⁽¹¹⁾. Stone-free rates are between 75% - 100% (5,12,13) which are compatible to those for kidneys with normal anatomy⁽¹⁵⁾. Stone-free rates are greatly increased with the use of auxiliary measures such as the flexible nephroscopy, multiple nephrostomy tracts and second-look procedures⁽¹²⁾. In our patient, PCNL was chosen because of the high stone burden. The first treated side was successful, rendering the patient stone-free. For the contralateral side, the stone clearance status for the remnant calyceal stones



Fig. 5 Unenhanced CT scan shows puncture of a right upper pole renal calyx that avoids the contents of an incisional hernia (arrows).



Fig. 6a Full length release IVU shows a left renal pelvic stone causing moderate obstruction in a malrotated kidney.



Fig. 6b Enhanced CT scan shows the spatial relationship of the kidney to the inferior right liver tip. Fluoroscopic-guided puncture of the left lower pole calyx is a safe approach in this case.

would have been enhanced had the patient agreed to secondary PCNL⁽¹⁶⁾.

In this era of minimally invasive surgery, the majority of renal stones can be successfully cleared with PCNL, ESWL or a combination of the two modalities. Open surgery for stone disease has become a rarity⁽¹⁷⁾. However, stone-bearing horseshoe kidneys may be one of its indications⁽¹⁴⁾. This can be done in conjunction with pyeloplasty or isthmectomy. In conclusion, while horseshoe kidneys are rare, the inherent poor drainage renders patients prone to infection, stone formation or even renal failure. Hence, maximal effort is required in order to conserve renal function, relieve obstruction, and achieve complete stone clearance.

ABSTRACT

Horseshoe kidney is the commonest congenital renal anomaly. Its inherently-poor drainage system renders the patient prone to stone formation. A 56-year-old man with bilateral renal stones in a horseshoe system associated with hydronephrosis is presented. He was treated successfully with bilateral CT-guided percutaneous nephrostomy and percutaneous nephrolithotripsy. Various treatment options, including ESWL, PCNL and open surgery, are discussed.

Keywords: horseshoe kidneys, renal caculi, extracorporeal shock wave lithotripsy (ESWL), percutaneous nephrolithotripsy (PCNL), intravenous urography (IVU)

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